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Lymphoplasmacytic NHL- A Rare Entity with Uncommon Presentation

Sanjay Marwaha, Nisha Marwaha, Parveen Malhotra*, Mahavir Jangra, Ritesh Kumar and Mehak Sharma
Department of Surgery, Pathology & Medical Gastroenterology, PGIMS, Rohtak, Haryana, India

***Corresponding author:**
Parveen Malhotra,
Department of Medical Gastroenterology, PGIMS, Rohtak

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1. Abstract

1.1. Back Ground

Non-Hodgkin's Lymphoma (NHL) presenting as an ileal mass, especially a primary ileal lymphoma, is rare but can occur, often involving the terminal ileum and caecum. Symptoms can include abdominal pain, altered bowel habits, weight loss, and a palpable mass in the right iliac fossa. Diagnosis is often confirmed through biopsy and histopathological examination.

1.2. Case Report

A thirty-one-year-old male patient, not a known case of any chronic illness, chronic smoker for last four years & chronic alcoholic for last four years but abstinent for last two months, presented with abdominal pain for last six months which was associated with intermittent diarrhea and blood in stools. There was weight loss of 4 kg in last six months. The ultrasonogram revealed mild ascites with thickened ileo-cecal junction with multiple mesenteric lymph nodes. The computed tomography scan showed asymmetric lobulated enhancing wall thickening involving terminal ileum, Ileo-cecal junction (ICJ), caecum, ascending colon with two polypoidal lesions in terminal ileum near ICJ, moderate ascites, mild hepato-splenomegaly with small left renal calculus. In view of above findings, patient was subjected to colonoscopy which showed grossly thickened bilateral ileal lips with few raised reddish mucosal, aphthae like spots in ascending colon. The ileal biopsy showed non-Hodgkin's lymphoma of lymphoplasmacytic type with CD 20 & 38 positive, BCl3 was focal positive, CD3 was sprinkling positive and CD19, Cyclin D1, lambda & Kappa were negative. The patient and family members were counselled for further investigations and treatment options but left against medical advice and were lost on follow up.

1.3. Conclusion

Lymphoplasmacytic NHL is rare and is a diagnostic challenge, especially when present with uncommon manifestations, as in our case in which it presented at younger age with selective involvement of Ileum without any lymphadenopathy.

2. Introduction

Non-Hodgkin's Lymphoma (NHL) presenting as an ileal mass, especially a primary ileal lymphoma, is rare but can occur, often involving the terminal ileum and caecum. Symptoms can include abdominal pain, altered bowel habits, weight loss, and a palpable mass in the right iliac fossa. Diagnosis is confirmed on biopsy and histopathological examination. Treatment may involve surgery, chemotherapy, or other therapies. Ileal NHL can involve the ileum as a primary site, or it can be a manifestation of lymphoma that has spread from another location. Abdominal pain, especially in the right iliac fossa, altered bowel habits (e.g., diarrhea or constipation), weight loss, and a palpable mass are common. Imaging (CT scan, etc.) can reveal the presence of a mass in the ileum. A biopsy of the mass is essential for definitive diagnosis, and immunohistochemistry can help determine the type of lymphoma. Treatment options include surgery (if the mass is resectable), chemotherapy, radiation therapy, and immunotherapy. The specific treatment approach will depend on the type of lymphoma, its stage, and the patient's overall health. The prognosis for ileal NHL varies depending on the specific subtype of lymphoma and the patient's overall condition. In differential diagnosis, conditions that can cause similar symptoms in the right iliac fossa, such as appendicitis, Crohn's disease, or other types of intestinal tumours, should be considered. NHL can also involve other organs, such as lymph nodes, bone marrow, and other parts of the gastrointestinal tract.

3. Case Report

A thirty-one-year-old male patient, not a known case of any chronic illness, chronic smoker for last four years & chronic alcoholic for last four years but abstinent for last two months, presented with abdominal pain for last six months which was associated with intermittent diarrhea and blood in stools. The pain was localized in right iliac fossa was intermittent and got relieved temporarily with symptomatic treatment including antispasmodics and analgesics. There were no aggravating or relieving factors. There

was no history of nausea, vomiting, fever, jaundice or constipation. There was weight loss of 4 kg in last six months. On physical examination, the patient was conscious, co-operative and afebrile. The systemic examination including chest, cardiovascular, central nervous system and ophthalmological was normal and there was no peripheral lymphadenopathy. The per abdominal examination revealed soft abdomen but no lump was appreciated. The complete hemogram revealed mild anemia with hemoglobin of 9.9 g/dL with normocytic normochromic picture, white blood cell counts & platelet counts were in normal range, with raised erythrocyte sedimentation rate (ESR) of 22. The renal function test, blood sugar, serum amylase & electrolytes, urine complete, thyroid & complete lipid profile, viral screen including hepatitis B, C, HIV, Electrocardiogram, chest x-ray were all essentially normal but liver function test showed hypoproteinemia and hypoalbunemia. The ultrasonogram revealed mild ascites with thickened ileo-cecal junction with multiple mesenteric lymph nodes. The computed tomography scan showed asymmetric lobulated enhancing wall thickening involving terminal ileum, Ileo-cecal junction (ICJ), caecum, ascending colon with two polypoidal lesions in terminal ileum near ICJ, moderate ascites, mild hepato-splenomegaly with small left renal calculus. In view of above findings, patient was subjected to colonoscopy which showed grossly thickened and ulcerated bilateral ileal lips with few raised reddish mucosal, aphthae like spots in ascending colon. The ileal biopsy showed non-Hodgkin's lymphoma of lymphoplasmacytic type with CD 20 & 38 positive, BCl3 was focal positive, CD3 was sprinkling positive and CD19, Cyclin D1, lambda & Kappa were negative. The patient and family members were counselled for treatment options but left against medical advice and were lost on follow up.

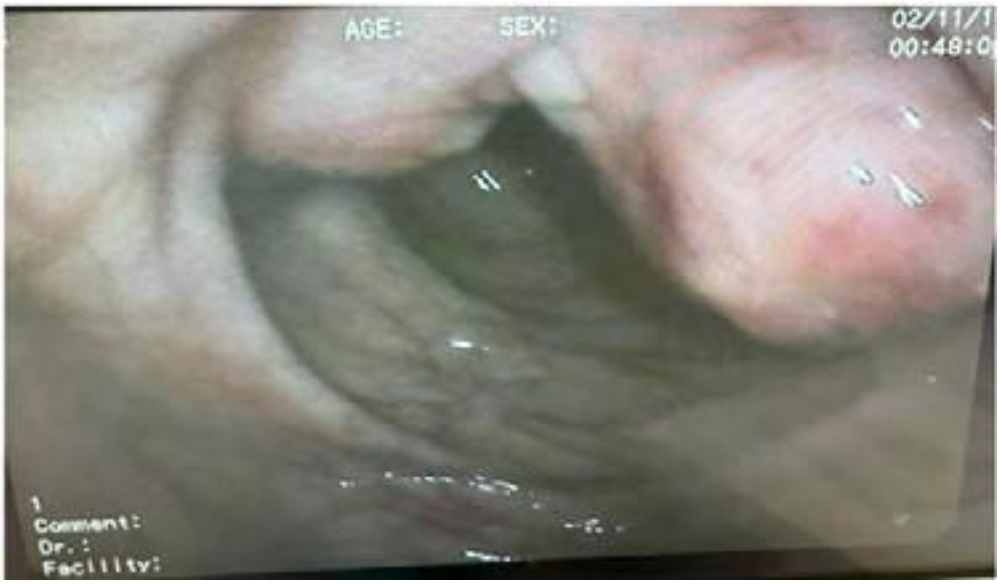

Figure 1: Showing Grossly thickened and Ulcerated Ileum.

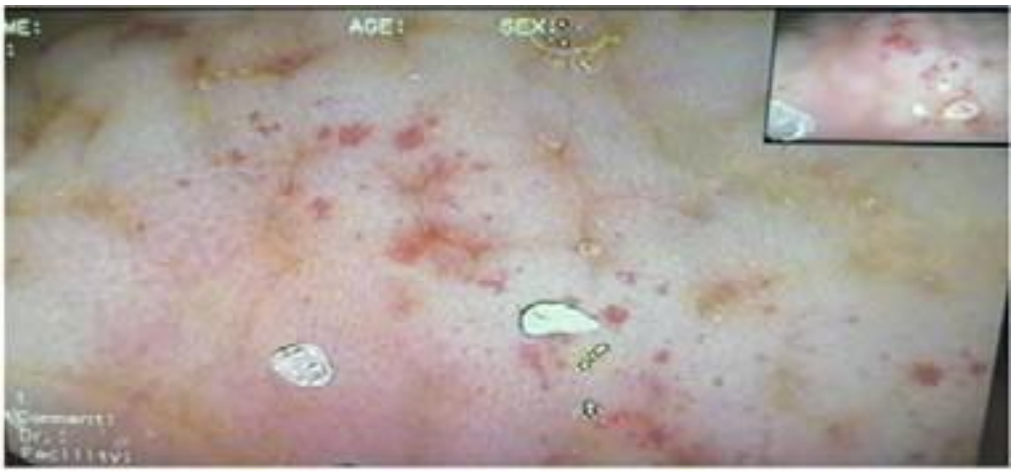

Figure 2: Showing Multiple Aphthae like Reddish spots in Ascending Colon.

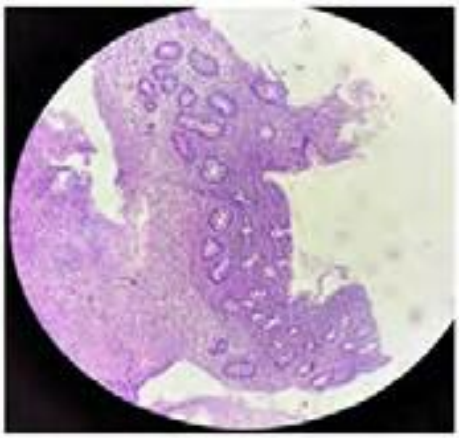

Figure 3: CD 20 Positive Specimen.

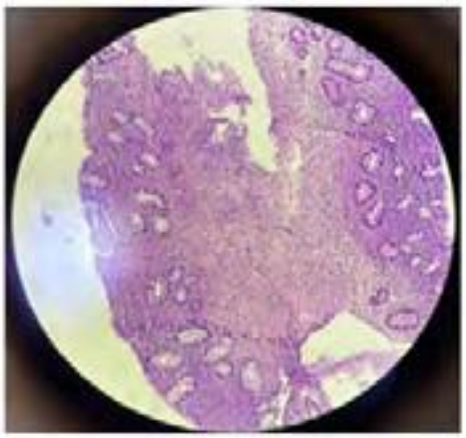

Figure 4: CD3 Positive Specimen.

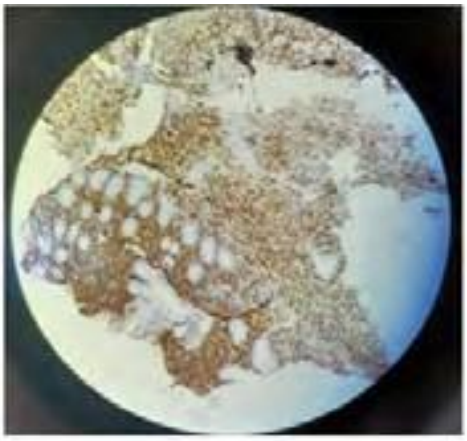

Figure 5: CD38+ Positive Specimen**.**

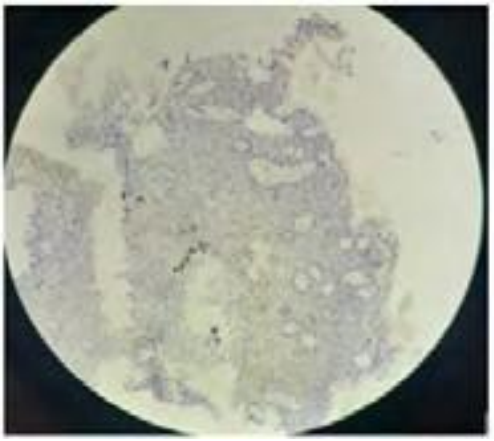

Figure 6: Cyclin D1 Negative Specimen.

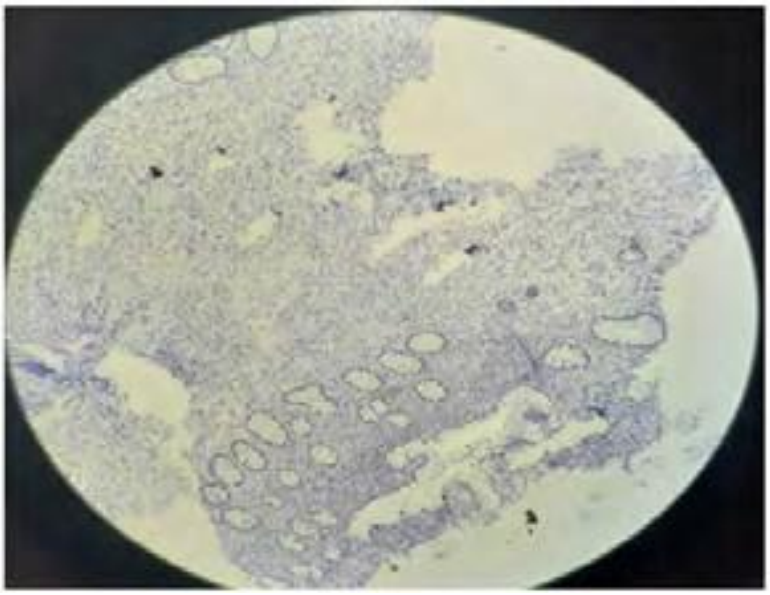

Figure 7: Kappa Lamda Negative Specimen.

4. Discussion

Non-Hodgkin lymphomas (NHLs) are tumors originating from lymphoid tissues, mainly of lymph nodes. These tumors may result from chromosomal translocations, infections, environmental factors, immunodeficiency states, and chronic inflammation. The clinical manifestations of NHL vary with such factors as the location of the lymphomatous process, the rate of tumor growth, and the function of the organ being compromised or displaced by the malignant process. The signs and symptoms of low-grade lymphomas include Peripheral adenopathy which is painless, slowly progressive and can spontaneously regress, primary extra nodal involvement and B symptoms and bone marrow involvement which associated with cytopenia's and fatigue/weakness. Intermediate- and high-grade lymphomas have a more varied clinical presentation like adenopathy, extra nodal involvement i.e. GI/GU tracts and B symptoms including Temperature >38°C, night sweats, weight loss >10% from baseline within 6 months; in approximately 30-40% of patients.Non-Hodgkin lymphoma (NHL) of the ileum is an intestinal tumor of intraepithelial T-lymphocytes, usually

presenting as a neoplasm composed of large lymphoid cells and is associated with necrosis and inflammatory background, as well as the presence of large numbers of histiocytes and eosinophils. Lymphoplasmacytic lymphoma (LPL) is a slow-growing, low-grade B-cell lymphoma, also known as Waldenström macroglobulinemia [1-4]. It's a type of NHL, which is a cancer that starts in white blood cells called lymphocytes, which are part of the immune system. LPL primarily affects bone marrow, where abnormal B lymphocytes and plasma cells accumulate. A significant number of LPL cases are associated with the production of an abnormal immunoglobulin M (IgM) antibody, often referred to as an IgM paraprotein. When LPL is associated with an IgM paraprotein, it's also referred to as Waldenström macroglobulinemia (WM). LPL is diagnosed through various tests, including blood tests, bone marrow biopsies, and imaging scans to assess the extent of bone marrow involvement. Treatment options for LPL depend on the stage and severity of the lymphoma, and may include chemotherapy, targeted therapies, and other treatments. LPL is a relatively rare type of NHL, affecting approximately 5 people per 1 million and can lead to a decrease in the production of healthy blood cells, potentially causing anaemia, neutropenia and thrombocytopenia. In some cases, LPL can lead to complications like hyper viscosity syndrome, where the blood thickens due to the accumulation of the IgM paraprotein. Lymphoplasmacytic lymphoma is a disease of elderly individuals in the seventh and eighth decade of life, with a slight male predominance. The disease is indolent, and most patients survive 7 to 8 years post-diagnosis. In rare cases, lymphoplasmacytic lymphoma can transform into an aggressive immunoblastic variant or other high-grade lymphomas [3-5]. As a rule, the diagnosis of lymphoplasmacytic lymphoma should be considered in elderly individuals with unexplained weakness, bleeding, neurological deficits, neuropathies, and visual difficulties [5-8]. Our case presented at a very young age of thirty-one which is in contrast to usual presentation in elderly group of lymphoplasmacytic non-Hodgkin's lymphoma. Moreover, there was no lymphadenopathy and prescence of B symptoms in the patient.

5. Conclusion

Lymphoplasmacytic NHL is rare and is a diagnostic challenge, especially when present with uncommon manifestations, as in our case in which it presented at younger age with selective involvement of Ileum without any lymphadenopathy. An extra vigil for uncommon and atypical presentations leads to timely diagnosis and proper management thereby decreasing morbidity as well as mortality associated with this life-threatening disease.

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